CASE REPORTS

Primary Spindle-Cell Sarcoma in the Right Auricle

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PRIMARY malignant tumors of the heart are rare, approximately 100 having been reported in the world literature to date.[12] Perlstein,[6] in an excellent review in 1918, collected reports of 31 cases and demonstrated the characteristics of the clinical and pathological syndrome produced by these tumors, namely, the multiplicity of symptoms and findings which do not fit the pattern of any common form of heart disease, and the frequent origin of these neoplasms in the right auricle. Bodenheimer,[2] who reported the first primary malignant cardiac tumor in 1865, well stated the clinical problem: "One can think of heart tumor when history, clinical examination, and observation do not speak for any other heart disease, when the symptoms occur rather suddenly and unexpectedly and the condition soon takes a faster, soon a slower course." Five cases of malignant cardiac tumors have been correctly diagnosed during life.[1, 7, 8, 9, 11] A few probable rhabdomyosarcomas have been reported.[1, 3, 4] Mahain[5] recently extensively surveyed the literature on all types of primary cardiac tumors and his studies of the sarcomas led him to conclusions similar to those previously mentioned.

CASE REPORT

The patient, a Mexican male 25 years of age, had been well until seven days previously. Then, while running to catch a streetcar, he noted for the first time an empty sensation in the epigastrium and slight shortness of breath. These symptoms persisted even without exercise, and three days later there developed substernal "heaviness" which seemed to ascend into the neck and which was associated with what the patient described as a "ball in the throat." The dyspnea and substernal oppression became progressively more severe. During the first five days following the onset of symptoms, the patient continued to work as a cement worker, but two days before admission to the hospital the symptoms became so severe that he was required to cease work. No other symptoms were elicited, and no further details referable to the present illness could be obtained from the patient.

Physical Examination: The temperature was 98.4° F., the pulse rate 130 and respirations 36 per minute. The blood pressure was 104 mm. of mercury systolic and 86 mm. diastolic in both arms. The patient was well developed, well nourished, alert, cooperative, mildly orthopneic, and apparently dyspneic.

Definite venous distention was noted in the neck. There was no tracheal shift and no enlargement of the thyroid gland.

The chest was symmetrical, with bilaterally equal respiratory movements, and there was no apparent deformity. On percussion, bilateral dullness was noted below the eighth thoracic spine posteriorly, and there appeared to be no descent of the diaphragm on deep inspiration. There was a suggestion of a change in dullness, with a shift to either lateral decubitus position. Breath sounds were diminished over this same area of dullness, with somewhat more diminution on the right side than on the left. Voice sounds were diminished over the same area. Moist inspiratory rales were noted over both bases; they were louder on the left.

The apex of the heart was approximately 2 to 3 cm. to the left of the midclavicular line in the sixth intercostal space, and dullness to percussion indicated that the right border was approximately 1 cm. lateral to the right sternal border in the fourth intercostal space. The heart sounds were very distant, and no murmurs, thrills, or rubs were noted. The rhythm was regular, and the pulmonic second sound was greater than the aortic second sound.

The liver was palpable 3 cm. below the right costal margin. The edge was smooth and the entire liver was slightly tender to deep palpation. No other organs were palpable, and no further abnormalities were noted.

Hemoglobin content of the blood was 14 gm. per 100 cc. Erythrocytes numbered 4.51 million and leukocytes 10,250 per cubic mm. Results of Wassermann and Kahn tests were positive. The packed cell volume was 40 mm. Corrected sedimentation rate (Wintrobe) was 0.0 mm. in one hour. Results of urinalysis were within normal limits.

Bilateral pleural effusions, with evidence of associated mediastinal or small pericardial effusion, were noted on an x-ray film of the chest (Figure 1).

Sinus tachycardia (120 per minute), with electrical alternans, was recorded upon an electrocardiogram. The tracing was consistent with that commonly observed in acute pericarditis.

Venous pressure was 380 mm. of water, and manual pressure over the liver caused the fluid in the manometer to go over the top (over 400 mm.).

Circulation time, arm to lung, was 10.5 seconds using 1 cc. of ether intravenously, and arm to tongue was 30 seconds using 6 cc. of sodium dehydrocholate (Decholin®).

The cardiac silhouette on a teleroentgenogram was of "water bottle" type and there was pronounced, generalized enlargement on fluoroscopy. Cardiac pulsations and vascular indentations were almost completely absent. In addition, the basal silhouette of the heart appeared broadened. The findings were consistent with clinical diagnosis of pericarditis with effusion.

Clinical Impression: (1) Possible rheumatic fever or other hyperergic disease with pleural, pericardial, and possibly peritoneal involvement. (2) Possible acute myocardial infarction.

The initial treatment consisted principally of oxygen in-

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halation, sedation, and salt restriction. On the second hospital day the patient rapidly became extremely dyspneic. The blood pressure was 120 mm. of mercury systolic and 85 mm. diastolic. A right thoracentesis was performed, and following the removal of 350 cc. of clear straw-colored fluid the patient fainted. Prompt recovery occurred after inhalation of ammonia fumes. Specific gravity of the pleural fluid was 1.013, and the total protein content was 2.3 gm. per 100 cc. Smears and cultures were negative for all organisms including Mycobacterium tuberculosis. An electrocardiogram showed sinus tachycardia (108 per minute) and the other changes which were consistent with acute pericarditis with effusion.

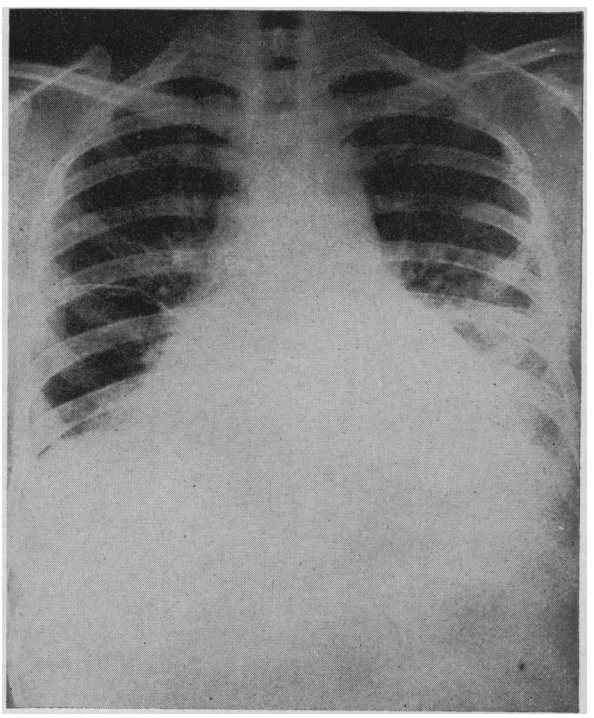

Figure 1.—X-ray film of chest.

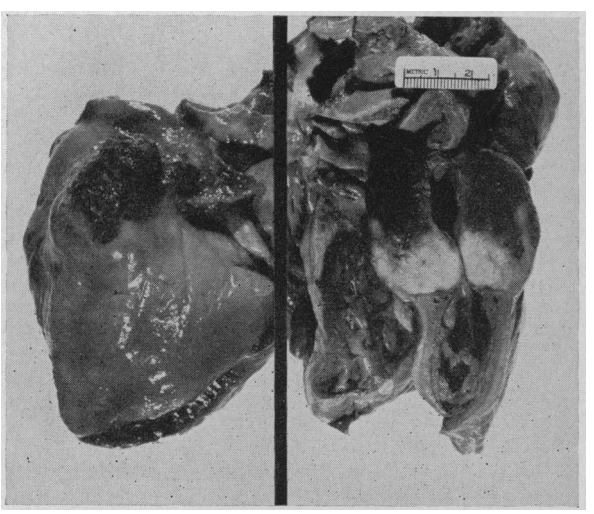

Figure 2.—*Left:* Neoplasm in right auricle with ulceration of its epicardial surface. *Right:* Hemisection through tumor showing hemorrhagic upper segment and the grayish yellow lower part with extension toward the insertion of the tricuspid valve.

On the fourth hospital day the patient became progressively more dyspneic. Left thoracentensis was performed, and 500 cc. of clear straw-colored fluid, with a specific gravity of 1.012 and a total protein content of 1.8 gm. per 100 cc., was removed. A right thoracentesis yielded 1,050 cc. of similar fluid. Later, left thoracentesis was repeated and 350 cc. of fluid was drawn. Mild syncope followed the procedure. At the conclusion the pulse rate was 86 per minute and the blood pressure 102 mm. of mercury systolic and 60 mm. diastolic. Although apprehensive, the patient was less dyspneic.

The patient died on the fifth hospital day after a night of progressively increasing dyspnea.

The patient was afebrile during his entire hospital stay.

At autopsy, seven hours after death, the only abnormalities noted on external examination were diffuse cervical edema typical of "bull neck" with distention of the cervical veins. No peripheral edema was present.

The pericardium was tautly distended with 1,150 cc. of bright red fluid having specific gravity of 1.030, which arose from the ulcerating, necrotic, deep red surface of a tumor mass in the body of the right auricle (Figure 1). The growth at its base measured 3 x 5 cm. and at its necrotic apex was 2 x 2.5 cm. The endocardium of the right auricle above the neoplasm was normal, but cutting 2 mm. into the endocardium of this area exposed the yellowish color of the tumor parenchyma. The right auricle was not dilated and the appendage was normal. The heart, with the growth, weighed 320 gm. Valve leaflets and chorda tendineae were normal. Valve circumferences were: Mitral 9 cm., aortic 7 cm., pulmonic 6.5 cm., and tricuspid 11 cm. The left ventricular wall measured 13 to 15 mm. and the right 2 to 5 mm. The chambers were not dilated. Coronary arteries were normal. Section through the long axis of the tumor showed it to be limited entirely to the auricular myocardium with extension to the insertion of the tricuspid valve (Figure 2). The tumor measured 3 x 5.5 cm. The inferior 2 x 3 cm. area of neoplasm consisted of grayish yellow material of firm consistency. The tissue above this was deep red in color and of mushy consistency. There was no evidence of metastasis of the tumor to other areas in the myocardium or to other organs.

Each pleural cavity contained 1,100 cc. of straw-colored fluid. The right lung weighed 1,050 gm. and the left 900 gm. The left lower lobe sank on immersion in water. Yellowish, foamy edematous fluid filled the tracheobronchial tree. The cut surface of the lung parenchyma was deep red in color, with rolled edges, and the surface still glistened after scraping. The lower border of the liver was at the costal margin and the organ weighed 1,400 gm. It was normal on the surface. On section it was noted to be grayish brown with increased lobular markings. No abnormalities were noted in gross and microscopic examinations of brain, meninges, thyroid, adrenal and pituitary glands, testes, entire gastrointestinal tract, pancreas, biliary system, kidneys, bone marrow, psoas muscle, rectus, and sternocleidomastoid muscles.

The parenchyma of the tumor consisted of moderately anaplastic pleomorphic spindle-shaped cells with deeply basophilic pleomorphic nuclei. The cells were loosely packed and tended to grow in parallel chains (Figure 3). Nuclei were not prominent. Lymphocytes infiltrated between some cords of neoplastic cells. In other areas clumps of lymphocytes were present. The growth at its margin invaded between normal muscle fibers. Scattered areas of fibrosis were present throughout. The central part contained areas of necrosis with polymorphonuclear and mononuclear infiltration. There were rare giant cells present which did not contain concentric striations. The upper part

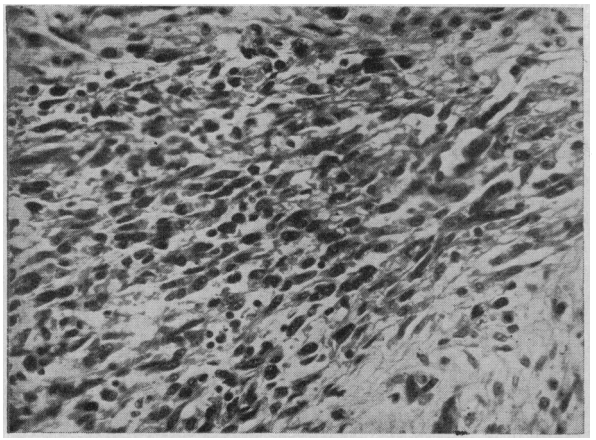

Figure 3.—Section through parenchyma of tumor showing spindle-shaped cells and fibrous stroma.

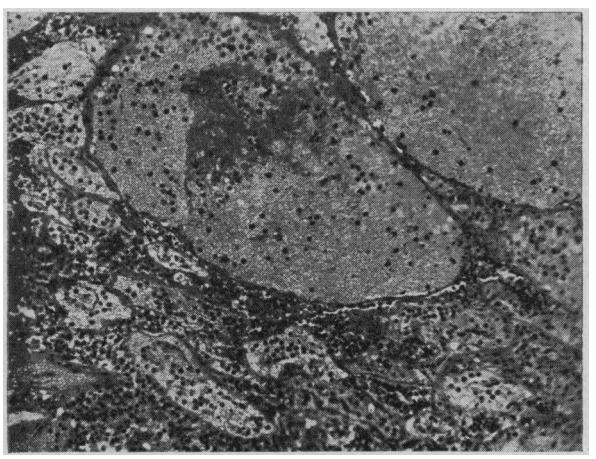

Figure 4.—Hemangiomatous upper section of tumor.

of the lesion was very vascular and resembled a hemangioma (Figure 4). Several tumor thrombi invaded the veins (Figure 5). With Von Gieson stains the tumor cells appeared yellowish pink and there was no connective tissue between them. There were moderate amounts of argentaphilic fibers about the tumor-cell bundles. No transverse striations were noted on Von Gieson stained specimens nor when hematoxylin and eosin, phosphotungstic acid and hematoxylin, or polaroid microscopy were used. Longitudinal fibrils were not demonstrated by phosphotungstic acid and hematoxylin stain. Patchy bronchopneumonia and pulmonary edema were noted upon section of the lungs. There was central congestion with atrophy of central liver cells and hemosiderin pigmentation in them.

DISCUSSION

Spindle-shaped cells such as made up the tumor parenchyma in this case are commonly noted in the reports of rhabdomyosarcoma of the heart, and giant cells are also common in this condition. The presence of tumor thrombus in the vein is characteristic of sarcomata. The giant cells in peripheral muscle rhabdomyosarcoma, however, have concentric striations, which were not found in the case here reported.

The Los Angeles Tumor Registry interpreted the neoplasm as being a "spindle-cell sarcoma in the right auricle."

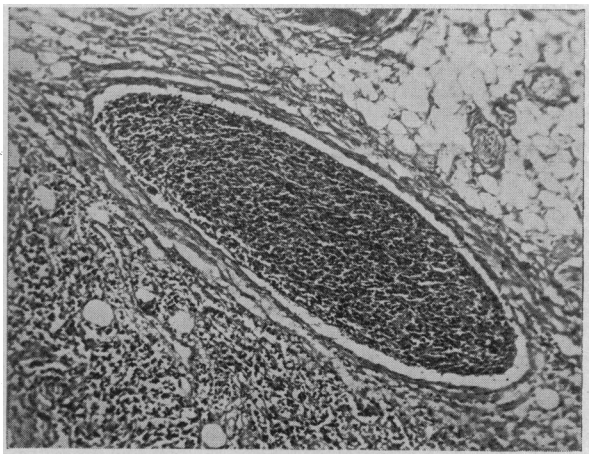

Figure 5.—Tumor thrombus in vein.

Concerning the diagnosis of rhabdomyosarcoma, MacCollum, as quoted by Larson and Sheppard,[3] stated very aptly: "Since the criterion demanded before an origin from muscular tissue will be admitted is the demonstration of characteristic cross-striations, it is not at all improbable; indeed it is almost certain that many tumors truly of such origin or character are not recognized as such, and are relegated to the tumor scrap heap of sarcoma."

SUMMARY

In a case of spindle-cell sarcoma in the right auricle there was some suggestion that the tumor was of muscle origin.

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